Limb reconstruction using circular frames in children and adolescents with spina bifida

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We report the outcome of 28 patients with spina bifida who between 1989 and 2006 underwent 43 lower extremity deformity corrections using the Ilizarov technique. The indications were a flexion deformity of the knee in 13 limbs, tibial rotational deformity in 11 and foot deformity in 19. The mean age at operation was 12.3 years (5.2 to 20.6). Patients had a mean of 1.6 previous operations (0 to 5) on the affected limb. The mean duration of treatment with a frame was 9.4 weeks (3 to 26) and the mean follow-up was 4.4 years (1 to 9). There were 12 problems (27.9%), five obstacles (11.6%) and 13 complications (30.2%) in the 43 procedures. Further operations were needed in seven patients. Three knees had significant recurrence of deformity. Two tibiae required further surgery for recurrence. All feet were plantigrade and braceable.

We conclude that the Ilizarov technique offers a refreshing approach to the complex lower-limb deformity in spina bifida.

In spina bifida the primary neurological lesion is in the lower motor neurone, leading to complex and variable combinations of flaccid paralysis and sensory loss. There may also be upper motor neurone lesions, leading to spasticity and other disorders of movement. These primary neurological lesions lead not only to impaired gait and function, but also to secondary musculoskeletal pathology, including contractures, bony torsional deformities, joint instability and degeneration. The management of lower-limb deformities is challenging because it is undertaken against the background of disordered gait, sensory impairment, muscle imbalance and osteopenia.

An individual patient’s lower-limb deformities cannot be predicted purely on the neurosegmental level and observed muscle imbalance or paralysis.1,2 Each child should be systematically examined for the presence of deformities affecting each segment of the lower limb and a clinical assessment of gait biomechanics should be undertaken. Three-dimensional (3D) gait analysis, albeit not essential, may help with preoperative planning. The aims of surgery can be tailored to realistic and achievable functional goals, ranging from the ability to wear a normal shoe, to facilitate appropriate lower-limb bracing or to improve independent walking in the high-functioning sacral level patient. Lower-limb deformities in spina bifida are often severe, sometimes rigid, technically difficult to correct, and have high rates of complications3-5 and risk of recurrence.6,7

The Ilizarov technique has been shown to be successful in the management of complex lower-limb deformities.8-15 However, there are no published series dealing specifically with the correction of these deformities in patients with spina bifida. Given the severity of deformities in this group, the high risk of complications with conventional surgery and the significant advantages afforded by a fine wire circular frame, it is perhaps surprising that this technique is not more widely accepted in these patients. We therefore undertook a retrospective cohort study to report our indications, complications and outcomes of the use of the Ilizarov technique in spina bifida.

Patients and Methods

Between 1989 and 2006, 33 patients with spina bifida underwent 48 corrections of lower limb deformities using the Ilizarov technique at three tertiary care children’s hospitals (The Royal Children’s Hospital, Melbourne, Australia; Musgrave Park Hospital, Belfast, United Kingdom; and Birmingham Children’s Hospital, United Kingdom). Five patients with less than one year of follow-up were excluded from this study, leaving 28 patients (15 boys and 13 girls) who underwent 43 deformity corrections (24 left, 19 right). A proforma was designed for data collection from the clinical
records and radiographs. Most patients had an annual standardised physical examination by a physiotherapist trained in methodology approved by the International Myelodysplasia Study Group, in which a comprehensive systematic examination includes assignment of the neurosegmental level. Three-dimensional gait analysis was used in Melbourne and Belfast, primarily as a research tool. Owing to the continuing development of this modality in analysing conditions such as spina bifida, its usefulness as a planning tool and outcome measure is still to be determined.

The surgical history before Ilizarov reconstruction, the indications for operation and the need for additional procedures after reconstruction were also recorded, as well as the use of orthoses, shoe modifications and the presence of pressure sores.

Complications were classified according to Paley, who considered difficulties during limb reconstruction as problems, obstacles or complications. A problem is a potential, expected difficulty that is fully resolved by the end of the distraction or fixation by non-operative means. An obstacle is a potential, expected difficulty arising during distraction or fixation and is fully resolved by the end of treatment by operative means. A complication includes any local or systemic intra- or peri-operative difficulty that remains unresolved at the end of treatment, and any early or late post-treatment difficulty. Complications were considered as minor or major and recurrent deformities and the need for further surgery during follow-up were also recorded.

The neurosegmental levels were thoracic in three, L1 in two, L3 in eight, L4 in six, L5 in five, and sacral in four. The 43 deformities corrected were divided into three groups for ease of description and analysis. Group 1 comprised 13 knee flexion deformities, group 2, 11 tibial deformities and group 3, 19 foot and ankle deformities.

The indications for surgery in group 1 depended on the functional ability and goals of the patient, physiotherapist and family. Community and household walkers with crouch gait secondary to a flexion contracture of the knee of > 30°, or between 20° and 30° with weak quadriceps, were offered surgical correction. In exercise walkers and non-ambulant children, surgery was indicated when the flexion deformity impeded sitting balance or the ability to transfer. In group 2, gradual derotation osteotomy was recommended for an external tibial rotational deformity > 40° which interfered with ambulation in higher-functioning patients, and the ability to transfer in the non-walkers. In group 3, surgery was indicated for any passively non-correctable complex foot deformity severe enough to preclude the use of an accommodative brace or shoe. Bony surgery was indicated on an individual basis, usually in children over eight years of age with more severe and rigid deformity. With evolution of the technique, fewer bony operations were performed and the mainstay of foot correction is now gradual soft-tissue correction with or without soft-tissue releases.

Results

A total of 16 patients had unilateral correction, eight had bilateral simultaneous correction and four had bilateral sequential correction. One patient who had bilateral simultaneous correction required a repeat correction on one side for recurrent deformity. One patient also had bilateral simultaneous corrections of his knees followed by correction of his tibiae.

In group 1 there were 12 contractures with a mean of 28.8° (20° to 45°) and one unstable neuropathic joint. The contractures all underwent frame-assisted soft-tissue distraction, unilateral in two patients and bilateral simultaneous in five. Seven knees underwent simultaneous posterior soft-tissue release, and the unstable neuropathic joint underwent arthrodesis (Fig. 1).

In group 2 there were seven primary external rotational deformities with a mean of 55.5° (40° to 80°) and one combination of external rotation and valgus. The contractures all underwent frame-assisted soft-tissue distraction, unilateral in two patients and bilateral simultaneous in five. Seven knees underwent simultaneous posterior soft-tissue release, and the unstable neuropathic joint underwent arthrodesis (Fig. 1).

In group 2 there were seven primary external rotational deformities with a mean of 55.5° (40° to 80°) and one combination of external rotation and valgus. Two patients underwent tibial lengthening and one underwent tibial lengthening combined with correction of valgus. In nine patients, derotations, angular corrections and lengthenings were performed with an Ilizarov-type circular frame, and two pure derotations were performed using the Taylor Spatial Frame (Smith and Nephew Inc., Memphis, Tennessee). Lengthening was performed in two patients with hemimyelodysplasia and discrepancies > 4 cm; lengthening of 4 cm to 6 cm was achieved. All lengthenings included an above-knee orthotic attachment to the frame to help maintain knee extension, and a foot piece for greater control.
Group 3 included seven planovalgus, two equinus, six equinovarus, two cavovarus and two equinocavovarus deformities. In eight feet a bony procedure or osteotomy was performed, including bilateral talectomy in a patient with severely deformed multiply operated feet; two lateral column lengthenings in planovalgus feet; and four subtalar fusions, two for cavovarus and two for planovalgus. In six feet distraction was facilitated by soft-tissue releases, and in another five the deformity was corrected without either bony surgery or soft-tissue releases.

The mean age of the patients at the time of Ilizarov treatment was 12.3 years (5.2 to 20.6). The mean age was 11.3 years (5.5 to 20) in group 1, 13.6 years (6.3 to 20.6) in group 2, and 12.1 years (5.2 to 20.3) in group 3.

In tibial deformities the construct was typically a three-ring frame fixed with a combination of 1.8 mm wires and half-pins. In foot deformities, the usual construct was two full tibial rings, with half-rings for the hindfoot and forefoot. The use of half-pins for transfixing the frame to the leg was much less common, as more experience was gained, because of their association with periprosthetic fractures in osteopenic bone. Towards the end of the series most of the frames were transfixed with fine wires alone (Fig. 2).

The children had undergone an overall mean of 1.6 previous operations (0 to 5) on the affected limb before Ilizarov correction, 1.1 (0 to 3) in group 1, 1.9 (0 to 5) in group 2, and 1.7 (0 to 3) in group 3. The earlier operations included soft-tissue releases, tendon transfers, osteoclast, osteotomy and arthrodesis. Recurrent deformities were numerous, and often the primary indication for the Ilizarov technique.

The mean duration of treatment in the circular frame was 9.4 weeks per procedure (3 to 26). In group 1, the mean was 9.5 weeks (6 to 26), in group 2 it was 12.1 weeks (5 to 18), and in group 3, 7.9 (3 to 15). Following removal of the frame, weight-bearing for six weeks in a plaster was followed by the application of a custom ankle-foot orthosis for long-term bracing. The short three-week frame correction in group 3 was for bilateral pure equinus deformity, and consisted of posterior releases followed by relatively rapid correction with the Ilizarov apparatus.

The mean follow-up for all patients was 4.4 years (1 to 9). It was 4.78 years (1 to 6.8) in group 1, 3.9 years (1 to 9) in group 2 and 4.4 years (1.7 to 8) in group 3.

There were 12 problems in the 43 procedures (27.9%, Table I). In group 1, one patient developed an ischial pressure ulcer within a week of operation. This resolved completely with local care before removal of the frame. Only two of the pin-site infections (group 3) required admission to hospital for intravenous antibiotics.

There were five obstacles in the 43 procedures (11.6%, Table I). In group 1 the patient with the arthrodesis of the knee lost fixation of her proximal ring and loss of position three weeks after application of the frame. She also developed a pressure sore from the ring. She underwent revision of the frame and achieved union at 26 weeks, with the pressure sore healing meanwhile.

There were 13 complications in the 43 procedures (30.2%, Table I). In group 1 there were five major complications in four patients. One sustained a supracondylar femoral fracture around a half-pin following minimal trauma. Re-operation was required to extend the frame to include the fracture, which healed without further complication. There were four recurrent flexion deformities in three patients. One developed bilateral deformities three years after removal of the frame and underwent bilateral supracondylar extension osteotomy. One patient who had bilateral simultaneous knee frames without soft-tissue releases underwent repeat frame application with soft-tissue releases, and had a successful outcome. The third patient who developed recurrent knee deformity required no further treatment.

There were five major complications in group 2. One patient had a severe refractory pin-site infection with osteomyelitis after removal of the frame and required drilling of the pin site, intravenous antibiotics and a vacuum-assisted closure dressing to heal her wound. She had no further recurrence of infection. There were two tibial fractures which occurred within a year of removal of the frame. One was sustained after a fall while rock climbing and was treated conservatively. The other occurred in a fall from an entrance ramp at school and was unrelated to the site of regenerate bone. This was also treated conservatively. Two patients developed recurrent tibial deformity.
There were three complications in group 3. The major complication was a pressure sore which occurred over the navicular after fitting of the ankle-foot orthosis. It healed after two years. The minor complications were a heel sore which resolved after adjustment of the ankle-foot orthosis, and persistent pain at the site of a transferred tibialis posterior tendon in another patient.

Further operations are either planned or have been performed in seven limbs (16.3%), five (11.6%) for recurrent deformity.

Two of the 11 limbs from group 2 have required another procedure for recurrent deformity. One had increasing external tibial rotation after lengthening, and the other was due to recurrent valgus of the proximal tibia. Both corrections were achieved with the Ilizarov frame. One patient had mild isolated ankle pain, but considered this to be better than before treatment of his tibial deformity.

In group 3, two of the 19 limbs are awaiting further procedures. In one patient, bilateral supramalleolar osteotomy is planned for external tibial torsion unrelated to the original Ilizarov correction of the foot. All the feet were recorded as plantigrade after treatment (Fig. 3). Although it is difficult to measure subtle changes in the flexibility of the hindfoot and midfoot in multiply operated feet, it was the author’s impression that, even in stiffer feet, some residual flexibility was retained after frame correction without fusion. All patients could wear normal shoes and were comfortable in their respective orthoses where appropriate. The patient with pain at the site of her transferred tibialis posterior tendon is being managed expectantly, and no patient had pressure sores at the latest follow-up.

### Discussion

Although there are limitations in reporting patients from three separate institutions, the authors share a common philosophy in the treatment of limb deformity in spina bifida. As this type of surgery is rarely performed in large numbers in this condition, the opportunity to report on this cohort has allowed new perspectives on the indications, complications and outcomes of the Ilizarov technique in spina bifida.

**Table I. Problems, obstacles and complications (after Paley17)**

<table>
<thead>
<tr>
<th>Group</th>
<th>Problems</th>
<th>Obstacles</th>
<th>Complications</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>One ischial pressure ulcer</td>
<td>One lost ring fixation: revised</td>
<td>Minor: One fracture around half-pin</td>
</tr>
<tr>
<td></td>
<td></td>
<td>One pressure ulcer from lost ring fixation</td>
<td>Major: Two tibial fractures</td>
</tr>
<tr>
<td></td>
<td></td>
<td>One broken wire: revised</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>Four pin-site infections</td>
<td>Two frame adjustments in the operating theatre</td>
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<tr>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>Seven pin-site infections</td>
<td></td>
<td>Minor: One pressure ulcer</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Major: One foot pain</td>
</tr>
</tbody>
</table>

**Fig. 3a**
Lateral radiographs of a) complex foot deformity pre-operatively and b) two years post-operatively.

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S. P. KELLEY, C. E. BACHE, H. KERR GRAHAM, L. T. DONNAN
The indications for surgery in group 1 were guided by the natural history. In higher-functioning children, those with knee flexion contracture > 20° are known to be much less likely to be able to walk in late adolescence. Treatment of knee flexion contractures by anterior hemi-epiphyseodesis is gaining popularity, but in our series most deformities were considered too severe and too late to be dealt with effectively by guided growth. At the time of the operations in our study, staples were the implant of choice for hemi-epiphyseodesis, and there were also concerns regarding reversibility in the younger child. New technologies such as lesion band plates and screws may promote anterior hemi-epiphyseodesis as a useful option in the management of knee flexion deformities in the younger child with spina bifida.

In group 2, gradual derotation osteotomy was recommended for an external tibial rotational deformity > 40° that interfered with walking in higher-functioning patients, and the ability to transfer in the non-walker. Tibial lengthening was considered for those higher-functioning children with a limb-length discrepancy > 4 cm which caused a significant functional disability. In group 3, surgery was indicated for any passively non-correctable complex foot deformity severe enough to preclude the use of an accommodative brace or shoe. The patients in this group were also considered unsuitable for a conventional surgical approach because of multiple previous operations or the severity of the deformity.

Although takedown has been a popular option for the severe, non-braceable, rigid equino varus foot after failed posteromedial release, it is a demanding operation with, at best, < 72% excellent and good results. It rapidly becomes an even less attractive option when one considers that it is not possible to correct severe deformity without further midfoot and forefoot osteotomies.

Soft-tissue procedures can be used to balance the foot with releases being preferable to transfers because of the unpredictable results of the latter. Fusion of the foot, in particular triple arthrodesis, has long been considered controversial. It is supported by Olney and Menelaus, but opposed by Dias, who believes that a rigid foot is more prone to pressure sores. However, triple arthrodesis may be considered for correction of deformity and has been associated with up to 83% satisfactory results. The majority of the patients in that series were community walkers with a sacral neurosegmental level and, by inference, would have deformities at the lesser end of the spectrum. Few reports of this approach are as successful, with one series showing a 33% failure rate. Patients with severe deformity are limited in the correction available from triple arthrodesis. The margin for error is particularly small for a patient with spina bifida. Any residual malalignment with the inherent stiffness of an arthrodesis can lead to trophic changes and pressure ulceration, and we think triple arthrodesis can no longer be recommended in spina bifida.

The Ilizarov technique has several advantages over conventional surgery. Plaster immobilisation is not required acutely, and therefore the associated complication of skin necrosis secondary to altered sensation and pressure areas should occur less frequently. Menelaus and Broughton reported a 61% incidence of pressure sores in a large cohort of patients with spina bifida, compared to our rate of 9.3% in 43 corrections. The ability to inspect the incisions and key pressure areas is reassuring to all, and we believe that this is a major step in reducing the morbidity associated with the conventional treatment of deformity in spina bifida.

In the osteopenic limb our preference is to use only purely fine wires for the frame, avoiding half-pins, which have a higher complication rate owing to cantilever loading and the development of a stress riser that can lead to fracture.

A gradual correction is desirable for several reasons. These limbs are scarred, and therefore have tenuous soft tissues and tethering of neurovascular structures, which are at risk with an acute correction. Gradual correction also allows fine-tuning in the corrective phase of treatment. Because of the unique deformities encountered, the co-operation of the patient in the final positioning of the limb to maximise their individual function is a distinct advantage. The use of a gradual non-fusion technique retains valuable flexibility of the hind and midfoot compared to an arthrodesis. Also, the Ilizarov technique allows minimal soft-tissue dissection, in accordance with good surgical technique.

The rotational deformities encountered can be particularly severe, measuring up to 80° of external tibial torsion in our series. This is too great for conventional osteotomy, but is feasible with the Ilizarov technique. The only acute complications encountered in a study using a distally based tibial osteotomy and acute correction in children with cerebral palsy were in those who had a correction > 40°. These complications were wound infection and excessive swelling. The fine wire frame is particularly useful when severe deformities predominate. It allows a greater range of corrective options, which is essential, as complete correction must be the goal. One must not be limited by a narrow range of surgical options, particularly as partial correction can lead to significant complications and further morbidity.

Leg lengthening has been described in patients with spinal dysraphism, despite earlier reservations about the wisdom of such a procedure. Haddad and Hill found that the patients appeared far more at ease with their frames and more able to mobilise independently than their counterparts undergoing lengthening for tibial or fibular reduction anomalies. Furthermore, there was no increase in the tibial healing index compared to other indications for lengthening.

A common concern with external fixators in the correction of paediatric deformity is that treatment can be painful. Haddad and Hill reported that their cohort of spinal dysraphism patients stood out from other patients undergoing leg lengthening in the ease with which their pain was controlled. It is also our experience that children with spina bifida tolerate this method of treatment extremely well.
owing to their relative freedom from pain during correction. This view is further supported by the rapid achievement of correction, minimising the time in the frame.

The disadvantages of the Ilizarov technique are those of any external fixation device. The most common complication is usually pin-site infection, with rates of up to 100% in many series following fine wire fixation.10-13 The problem is dealt with relatively easily by local care, including oral antibiotic treatment in the majority of cases. We have not seen an increase in pin-site infection in spina bifida patients more than would be expected in our regular limb reconstruction patients who have Ilizarov frames.

In conclusion, we have found that the Ilizarov technique offers a refreshing approach to complex lower-limb deformity in patients with spina bifida.

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No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.

References


